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CONTROVERSIES

Nancy L. Snyderman, MD, and Richard J. H. Smith, MD, *Editors*

FIBROUS DYSPLASIA

Consultants: Donald Kearns, MD, Trevor McGill, MD, and William Potsic, MD

When faced with a benign but destructive neoplastic process in an infant, the obvious questions raised are what to do about it and when. That's precisely the dilemma posed to our experts.

A 12-month-old boy was noted to have a slight swelling over the right malar eminence. By the age of 18 months the deformity was easily noticeable. In addition to the swelling, epiphora developed secondary to obstruction of the right nasolacrimal duct. The overlying skin was freely mobile. There was no history of epistaxis and no obvious discomfort. Radiographs were obtained (Figures 1 and 2).

1. Based on this child's history and the radiographic findings, what are your considerations?

Dr. Kearns: My differential diagnosis would include fibrous dysplasia, ossifying fibroma, maxillary sinus osteoma, fibrous sarcoma, aneurysmal bone cyst, giant cell tumor, or an odontogenic tumor.

Dr. McGill: Considerations include fibrous dysplasia, ossifying fibroma, osteoochondroma, osteoblastoma, and cementifying fibroma. However, based on the clinical history and computed tomographic (CT) scan, the most likely diagnosis is fibrous dysplasia.

Dr. Potsic: The history and radiographic findings indicate a fibro-osseous lesion. The spectrum ranges from fibrous dysplasia to malignancies of osseous origin, and all must be considered. In this case I suspect the diagnosis is fibrous dysplasia.

2. After making the diagnosis of fibrous dysplasia, would you recommend additional diagnostic tests?

Dr. Kearns: A bone scan would be positive if the disease is active. It is a dependable way to follow the dysplasia and determine when the inactive phase is reached.

Dr. McGill: When the diagnosis of fibrous dysplasia is made, other skeletal lesions should be searched for and evaluated by a radiographic survey or bone scan. The patient should also be examined for pigmentary lesions and signs of precocious puberty. These steps are important to rule out polyostotic fibrous dysplasia. A CT scan will distinguish between the distinct clear mar-

Readers are invited to submit particularly difficult cases for consideration to Nancy L. Snyderman, MD, Editor, Controversies, ***Head & Neck,*** *Department of Otolaryngology–Head and Neck Surgery, California–Pacific Medical Center, 2100 Webster Street, Suite 202, San Francisco, CA 94115.*

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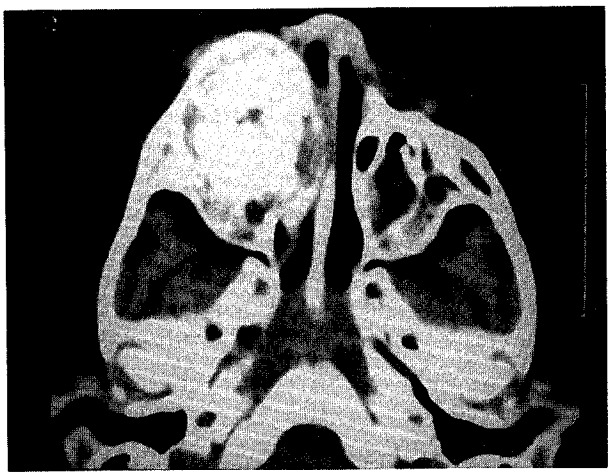

FIGURE 1. CT scan revealing a mass of the right malar eminence.

gins of ossifying fibroma and the ill-defined margins of fibrous dysplasia.

Dr. Potsic: After making a diagnosis of fibrous dysplasia I would recommend magnetic resonance imaging (MRI) and a MRI angiogram to determine the vascularity of the lesion and identify feeding vessels. This is very important before planning surgery. I would also obtain a bone survey, blood urea nitrogen (BUN), calcium, and phosphorous to eliminate concerns about polyostotic fibrous dysplasia.

3. Based on the sites of involvement, what are your immediate concerns?

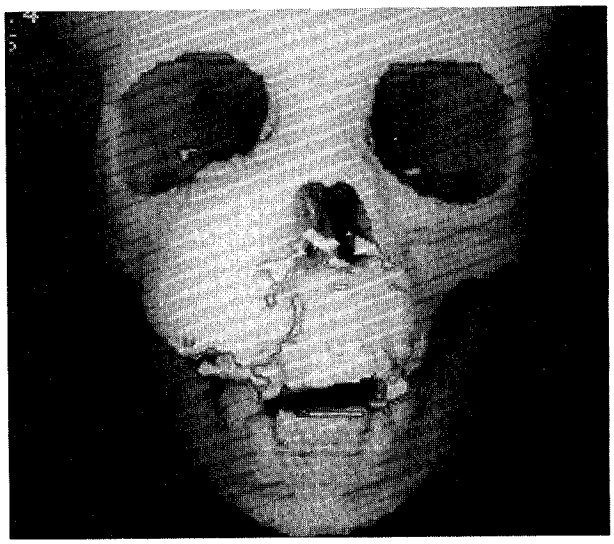

FIGURE 2. Three-dimensional reconstruction of the facial skeleton and tumor.

Dr. Kearns: My immediate concerns are impingement superiorly into the orbit and compression of the optic nerve. I also would be worried about malocclusion, erosion of permanent teeth, nasal obstruction, and sinusitis.

Dr. McGill: Immediate concerns are both cosmetic and functional. This tumor has already compromised the function of the nasolacrimal apparatus. Sudden growth in this area will likely cause a significant cosmetic defect. In addition, extension into the orbit is possible. The functional problems of nasolacrimal obstruction and proptosis will likely require early surgical intervention.

Dr. Potsic: Based on the site of involvement my immediate concern would be the activity of the fibroosseous lesion. If it is rapidly growing, there may be further nasolacrimal duct obstruction, orbital or ophthalmic complications, and altered cosmesis.

4. When would you recommend resection?

Dr. Kearns: I would recommend surgery for orbital decompression when early signs of decreasing visual acuity are noted. Otherwise, if the tumor is in a slow-growing phase, surgery may be elective.

Dr. McGill: I would recommend early surgery if functional problems, such as nasolacrimal obstruction and proptosis, are present. Otherwise, I would delay surgery until the dysplastic site has matured and is replaced by lamellar bone.

Dr. Potsic: Because this lesion is benign, an aggressive approach is not required at this time. Removal by locally excising the lesion should be sufficient if functional problems are present.

5. How aggressive would you be in trying to prevent proptosis or exophthalmos?

Dr. Kearns: I would not do prophylactic surgery to prevent proptosis or exophthalmos. Rather I would follow the patient carefully with good serial ophthalmologic exams. When proptosis occurs and shows signs of progressing I would decompress the orbit using techniques described for Graves' ophthalmopathy.

Dr. McGill: Extension of fibrous dysplasia into the orbit would be extremely rare. However, if this occurs, a complete surgical resection rather than a contouring procedure whould be performed to decompress the eye.

Dr. Potsic: Because this lesion is benign, an aggressive approach is not required. Local excision of the lesion should be sufficient. The lesion does not appear dense and should lend itself well to curettage.

6. What surgical approach would you consider when planning definitive treatment?

Dr. Kearns: A midface degloving or a lateral rhinotomy approach can be used to resect the lesion. I prefer the former because facial scarring is avoided.

Dr. McGill: A time-saving measure might be functional endoscopic decompression of the nasolacrimal apparatus. However, this technique would need to be repeated several times. The most definitive treatment would be complete resection via a midface degloving approach. This technique provides excellent exposure.

Dr. Potsic: I would prefer a Caldwell-luc or lateral rhinotomy approach. I favor the lateral rhinotomy because it provides maximal exposure. Although it has a cosmetic disadvantage, I believe it is the best approach.

7. Postoperatively, how would you follow this patient for recurrence of disease?

Dr. Kearns: I would follow this patient clinically with routine ophthalmologic exams to assess visual acuity and proptosis. When growth appears to be minimal, a bone scan may be used to determine whether the disease is in an inactive phase, thus allowing for elective cosmetic surgery. CT evaluation may also help.

Dr. McGill: I would follow this patient with examinations every 6 months and a CT scan on an annual basis.

Dr. Potsic: I would follow this patient with interval physical examinations and assess any functional defect. If the fibrous dysplasia is not active and not causing a progressive functional deficit, no additional intervention would be required. Serial CT scans should be helpful to assess the activity of the lesion.

8. Are you concerned about sarcomatous changes with this lesion?

Dr. Kearns: Sarcomatous degeneration is rare in fibrous dysplasia and is more common in the polyostotic type than in the monoostotic type. Sarcomatous degeneration can occur following radiotherapy. If that occurs, the tumor is usually an osteosarcoma rather than fibrosarcoma or chondrosarcoma.

Dr. McGill: Malignant change in fibrous dysplasia has only been described in patients with the polyostotic form of the disease and in people who undergo radiotherapy. Sarcomatous degeneration should not be a concern for this patient.

Dr. Potsic: Malignant degeneration of fibrous dysplasia is not a significant concern for this child. Radiotherapy is a risk factor for sarcomatous change but it would not be used in this case.

SUMMARY

The controversy is this case centers around the management of this lesion, not the differential diagnosis. All the consultants agreed that the history, physical, and CT findings were consistent with various fibroosseous lesions, the most likely being fibrous dysplasia.

The need for additional tests varied with a bone scan (Dr. Kearns), a bone scan and CT scan (Dr. McGill), and MRI, MRI angiogram, bone survey, BUN, creatinine, calcium, and phosphorus (Dr. Potsic).

Cosmetic and functional changes were considered priorities for the consultants, with orbital compression, malocclusion, tooth eruption, nasal obstruction, and sinusitis (Dr. Kearns), nasolacrimal duct obstruction and orbital compression (Drs. McGill and Potsic) being the concerns.

Because this lesion is benign and slow-growing, the consensus is that surgery should be reserved for functional or cosmetic compromise. But how agressive should one be and what approach should be used? The approaches varied with midface degloving or lateral rhinotomy (Dr. Kearns), midface degloving (Dr. McGill), or a Caldwell-luc and lateral rhinotomy (Dr. Potsic).

Assessment of this tumor postoperatively should be with patient examinations and serial CT scans. None of the consultants worried about sarcomatous changes in this tumor.